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Capillary haemangiomatosis: a rare cause of pulmonary hypertension

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Introduction

Capillary haemangiomatosis is a very rare cause of pulmonary hypertension with only 16 cases to date cited in the literature (1–5). The histological features are those of gross proliferation of small, thin-walled vessels infiltrating the lung parenchyma and walls of the larger vessels, particularly veins (6).

We report a case in a lady having previously undergone pneumonectomy.

Case Report

A 60-year-old single lady presented with increasing breathlessness on exertion, orthopnoea and swelling of her ankles. She had undergone right pneumonectomy in 1952 (aged 26) for removal of a pulmonary dermoid cyst. On examination she was dyspnoeic at rest and centrally cyanosed. The jugular venous pressure was elevated to the angle of the jaw and there was oedema of both legs. The heart rhythm was atrial fibrillation with an apical rate of 110, her left lung had basal inspiratory crackles.

Investigations showed a haemoglobin of 14·3 g dl^{-1}, haematocrit 47%; electrolytes, calcium, liver function and serum thyroxine were within the laboratory reference range. The chest radiograph revealed the right pneumonectomy but with a central mediastinum. The left lung was plethoric, with no evidence of upper lobe blood diversion or pulmonary oedema. ECG confirmed atrial fibrillation and right ventricular hypertrophy. The echocardiogram demonstrated large right chambers with mild tricuspid regurgitation; left ventricular function was good, and no left-to-right shunt was demonstrated.

The presentation was one of hypoxic pulmonary heart disease, although the cause of the presumed pulmonary hypertension was not evident. She was treated with oral diuretics (frusemide 80 mg + amiloride 5 mg daily) and at 1 month was clinically much improved with a satisfactory exercise tolerance, now managing her housework. A week later she deteriorated with increasing dyspnoea and oedema despite diuretics. An ACE inhibitor (captopril 25 mg BD) was introduced but made little difference. A lung ventilation:perfusion scan showed no significant perfusion defects in the left lung. She remained severely hypoxic (blood gases on air: PCO_2 8·5 kPa, PO_2 4·4 kPa) and oxygen therapy resulted in CO_2 accumulation (blood gases on 24% oxygen: PCO_2 10·8 kPa, PO_2 5·0 kPa). Spirometry showed a severe ventilatory defect with a FVC of 0·41 and FEV 0·31. A limited CT scan of her thorax demonstrated only pulmonary plethora in the remaining left lung with no evidence of interstitial lung disease. Right heart catherization confirmed severe pulmonary hypertension with right atrial pressures of 31/26, right ventricular pressures of 114/22 and pulmonary outflow tract 121/50, a mean of 84 mmHg. She died 2 months after her initial presentation.

Autopsy examination revealed mild hypertrophy of the right ventricular wall; coronary arteries and heart valves were normal. Macroscopically the left lung appeared normal, although subsequent histological analysis demonstrated diffuse capillary proliferation with a 'double layer' of capillaries on either side of the alveolar wall and capillary infiltration of the major arteries and veins with thrombosis of the pulmonary arteries as illustrated (Plate 1). These are the features of capillary haemangiomatosis.

Discussion

Capillary haemangiomatosis was originally described in 1978 (6). It is mainly detected in young adults (mean age 35 years; range 14–71) (1–3). The aetiology is unknown, although familial cases with apparent autosomal recessive inheritance (7) or congenital cases (8) are reported. Capillary haemangiomatosis has been regarded as a vascular tumour of

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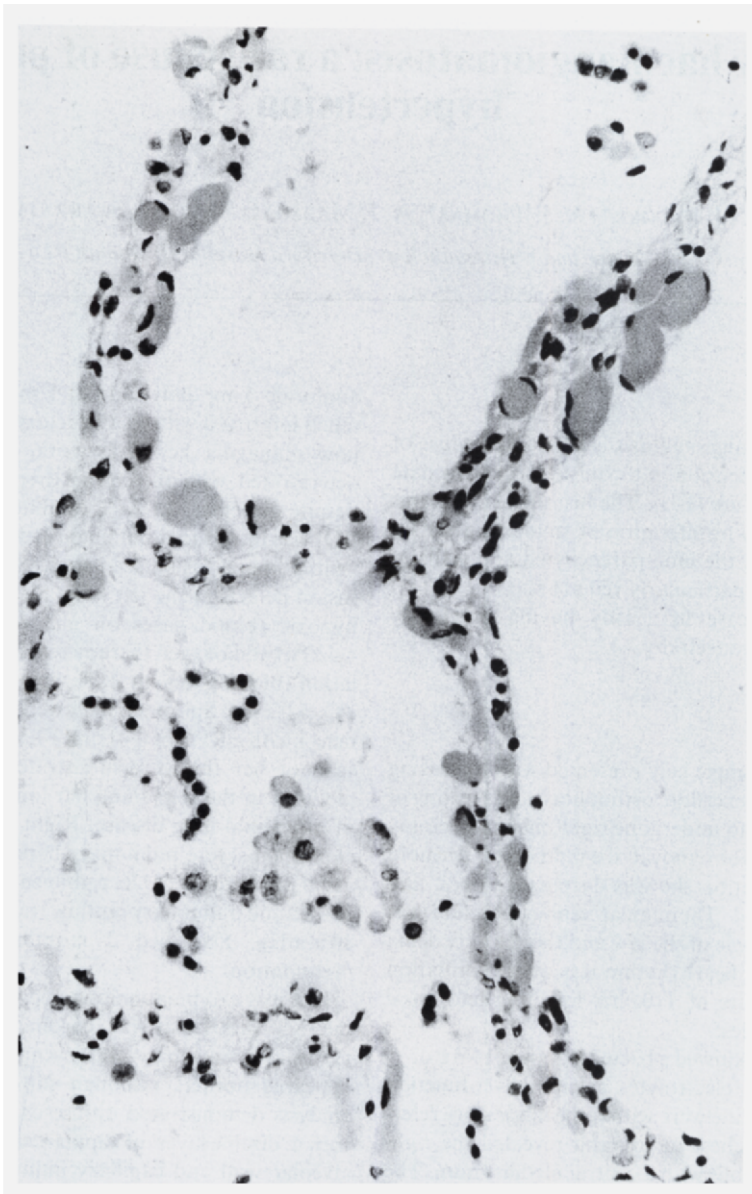

Plate 1 Histological section showing double layer of capillaries on either side of alveolar membrane, pathognomonic of capillary haemangiomatosis. Haematoxylin and eosin stain; × 800.

low-grade malignancy, where capillary vessels can increase greatly in size (9).

Pneumonectomy *per se* is not regarded as a sufficient cause of major pulmonary hypertension (10). The formal diagnosis was not made until autopsy, with none of the previous investigations providing diagnostic clues as had been reported by other authors (4, 5). There was no history of haemoptysis which can be a presenting symptom (3), although a relatively uncommon feature of pulmonary hypertension (11). In this case neither the chest X-ray nor the thoracic CT scan suggested interstitial lung disease, whereas most reports indicate a generalized increase in lung markings in a diffuse reticulo-nodular pattern on the chest radiograph in keeping with the nodular proliferation of capillaries (3). Varying degrees of cardiomegaly and enlargement of the pulmonary arteries are also noted consistent with measures of elevated

pulmonary arterial pressure. Pulmonary thrombosis with abnormal perfusion scans is usually also documented, although, as in our case, the $V:Q$ scan may be negative (3).

Spirometry indicated that there was a severe ventilatory defect, which is not a recognized feature of this condition, as judged by other reports (3, 4). At this stage our patient was extremely ill, and it is likely that this recording did not reflect the true physiological state. It is probable that the previous pneumonectomy contributed to the cor pulmonale. Re-examination of the histology of the 1952 pneumonectomy specimen in this patient has confirmed that the right lung contained a benign cystic teratoma containing skin appendages, benign glandular tissue and thymic gland tissue. There was no evidence of vascular changes attributable to pulmonary hypertension.

Treatment of this condition, as with other forms of pulmonary hypertension, is problematic. The use of vasodilators in primary pulmonary hypertension is disappointing, these agents being of prognostic but not of long-term therapeutic value (12). No specific therapy for capillary haemangiomatosis is described, although successful use of recombinant interferon has been reported (5). Interferon α-2a produced a beneficial response in a 12-year-old boy over a 14-month period with resolution of exertional dyspnoea and clubbing, normalization of the pulmonary pressure–volume curve, and a regression of the abnormal vascular pattern seen on pulmonary angiography (5). It is suggested that interferon inhibits proliferation of endothelial and smooth muscle cells. Long-term oxygen therapy does not appear to have been mentioned in reports as a possible therapeutic aid. Orthotropic heart-lung or single or double lung transplantation is likely to remain the only therapeutic hope for the foreseeable future (3).

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